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(9)

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A CASE OF "ANGEIO-KERATOMA."

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IT is twenty years ago since the disease now known as Angeio-keratoma was first described by Cottle in the *St. George's Hospital Reports* (1878), but the record long escaped notice. A case seen by Crocker in 1885 is mentioned in his work on "Diseases of the Skin." In 1886 and 1889 a number of cases were published independently by Colcott Fox. In 1889 the anatomical seat of the condition was recognized by Mibelli, and further careful and minute observations were added by Pringle in 1891. Since this time many cases have been reported by Dubreuilh, Tommasoli, Thibierge, Audry, Joseph, Fordyce,† and others.

The disease may be described as a multiple capillary angeiectasis, tending to the formation of small tumour-like prominences under the epidermis. It is almost invariably localised to the hands or feet, or both, but occasionally invading other parts of the body. In the hands and feet it is nearly always associated with a tendency to chilblains, and sometimes with more or less local asphyxia, and the superjacent epidermis undergoes a verrucose thickening (whence the name "keratoma"), but the cuticular hypertrophy is absent when the growths affect other parts of the body, and is, therefore, not an essential part of the complaint. It appears to be equally common in

* The case was shown at the Dermatological Society of London in December last, but the notes were held over until they could be published in full.

† *Journal of Cutaneous and Genito-Urinary Diseases*, N.Y., March, 1896. Full bibliographical references will be found in this article.

the two sexes, and first shows itself in childhood or adolescence, usually in weakly subjects, and after progressing for a variable number of years, tends to become stationary during, or even before, adult life. Spontaneous retrogression or disappearance has not yet been observed, except, perhaps, in one of the cases published by Dr. Colcott Fox, in which the spots, according to the statement of the friends, vanished completely during the last stages of a fatal illness. The disease is seldom attended by pain, pruritus, or other subjective symptoms.

In many cases it affects more than one member of the family, but there is at present no example of transmission by inheritance.

The peculiarity of the present case lies in its widespread distribution, in the almost complete immunity of the hands and feet, and in the absence of any tendency to chilblains or local embarrassment of circulation. The condition had given the patient so little trouble that he did not think it necessary to ask medical advice until he became inconvenienced by the appearance of a neuroma on the thigh. The association with a congenital deformity of the hands may probably be regarded as a mere coincidence, although an allied condition, a congenital contraction of the little finger, was noticed in one of the cases published by Dubreuilh. An attack of rectal hæmorrhage and a trace of albumen found in the urine when the patient first appeared in the out-patient room, may be mentioned as possibly related to the diseased condition of vessels. The only recorded case bearing any resemblance to this is that of Fordyce (*l. c.*), in which the scrotum was the seat of the affection.

The term "Angeio-keratoma" is unsatisfactory, in view of the recent development of our knowledge. The warty condition found in cases where the disease is limited to the hands and feet appears to depend upon purely local causes, and is slight or altogether absent when the affection occurs in other parts of the body, and the vascular lesion is not a new formation, as implied by the suffix "oma," but an ectasia of the cutaneous capillaries. The name proposed by Thibierge, "Acro-tele-angeiectasia," is more formidable but more scientific, and might be adopted till a better presents itself.

The notes of the case are as follows :—

The patient, a house-painter, aged 39, was admitted into St. Thomas's Hospital in December last, suffering from neuroma of the

thigh, multiple micro-angeiomata of the trunk and extremities, and a congenital deformity of the hands.

History.—The man had suffered from typhoid fever during his boyhood, and at the age of 18 from an attack of bleeding from the rectum, lasting about five weeks. Apart from these illnesses he has always enjoyed good health. He has never been subject to chilblains or undue coldness of hands or feet.

The skin lesions were first noticed at the age of 11, and appeared

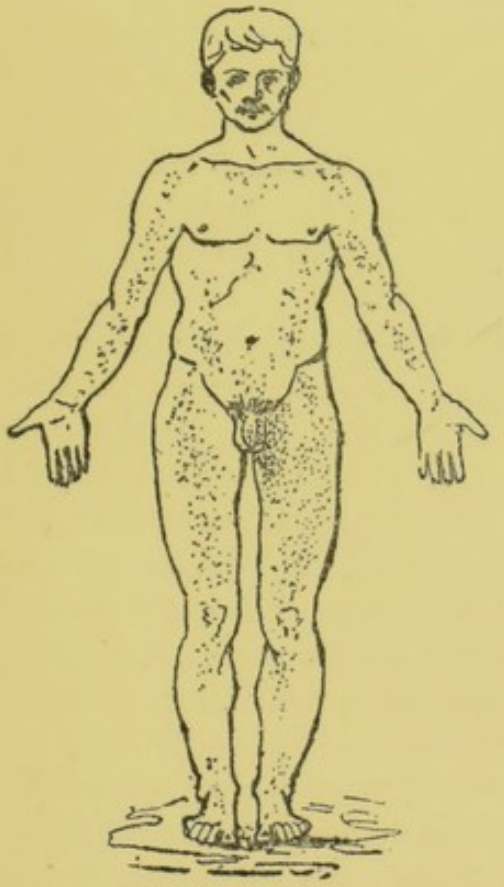

FIG. 1.—Distribution of the Lesions over the Front of the Body and Limbs.

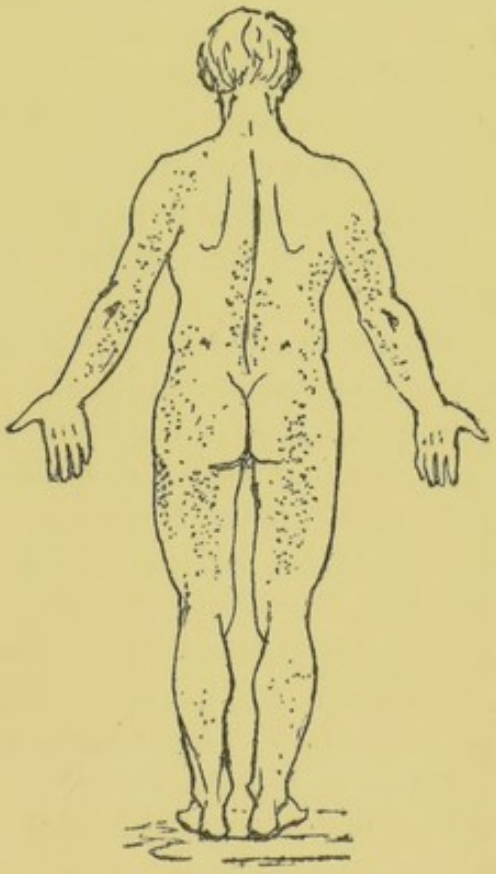

FIG. 2.—Posterior View.

in front of the knees, gradually spreading to the trunk and upper extremities, and reaching their present extent about seventeen years ago. They have caused him no trouble, and he has not applied for treatment.

The neuroma appeared about three years ago. Some varicosity of the veins of the lower extremities was noticed about the same time, but has not given rise to any inconvenience. Family history is

negative with regard to angeiomata, but his mother and sister, and three out of four of his own children, had congenital deformities like his own.

Present condition.—The patient is somewhat under middle height, well-made, and muscular. His hair and eyes are dark, complexion healthy, but with slight dilatation of cutaneous capillaries. Urine normal since admission, but a trace of albumen was found at the time of application. The fingers of both hands are contracted at the middle and distal joints, and the middle and distal phalanges of the fourth finger on each hand are duplicated, the two digits being enclosed in one cutaneous investment. On the outer side of the right thigh, a little behind and below the great trochanter, is a subcutaneous neuroma, of the size and shape of a small kidney-bean, apparently connected with branches of the lesser sciatic nerve, and on pressure giving rise to pain extending down the back of the thigh.

The angeiectases appear as innumerable puncta and papules of a purplish-red colour dispersed over the whole surface except the face, palms, and soles (Figs. 1 and 2). They range in size from a mere point to that of a hemp-seed, reaching the greatest development on the scrotum, and on the inner side of the left thigh, and are smallest and least numerous over the hands, feet, and thorax. The largest form hemispherical projections, purple in colour, soft in consistence, emptying partially under pressure, and presenting no verrucose thickening of the cuticle; while the smallest are level with the surface, impalpable, and look like flea-bites. The larger spots are the most closely clustered, but show no tendency to fuse into patches, or to become grouped into definite figures of any kind, and the intervening skin is normal, except for a few enlarged vessels here and there. The larger angeiectases yield a drop or two of blood on pricking, but their prominence is not visibly diminished. There is no pruritus.

On December 13th the neuroma, a firm fibrous growth, was excised, and a small piece of the angeiectasic skin on the inner side of the left thigh was removed for examination.

On section of the affected integument the red papules are found to present a varicose dilatation of the capillaries of the papillary layer of the derm, the enlargement of the lumen of the vessel reaching its maximum near the membrana propria, there assuming a more or less

spheroidal form, and leading to a thinning, sometimes going on to almost complete disappearance, of the superjacent Malpighian layer of the epidermis, while the cuticle is either attenuated or remains unchanged. The interpapillary cones of the Malpighian layer around the vascular dilation are somewhat hypertrophied, and in places seem to completely surround the ectasia, an appearance probably due to the section having passed through the periphery of a spherical expansion. The walls of the varicosities are formed by a single layer of flat nucleated cells, and the lumina are occupied by blood coagula. Some of the larger vascular spaces are apparently

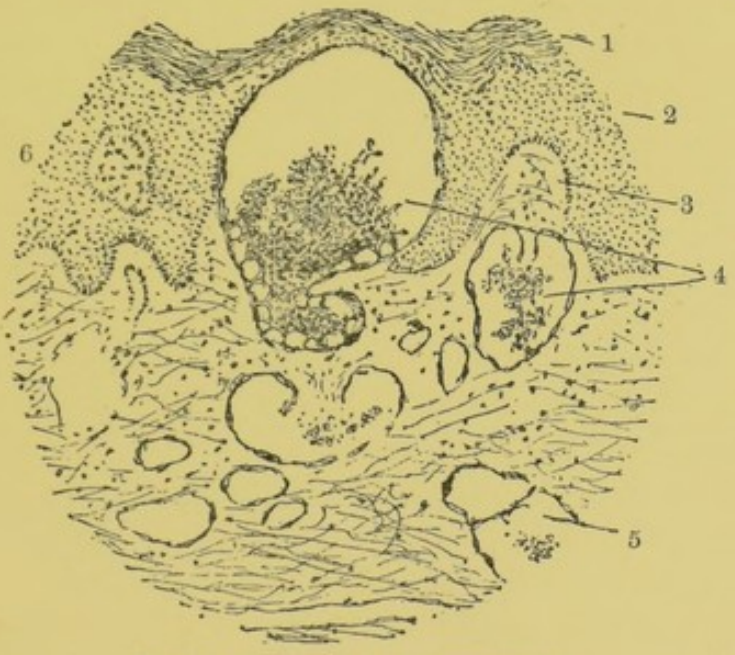

FIG. 3.—SECTION OF ANGEIECTASES.

1, Stratum Corneum of Epidermis; 2, Stratum Malpighi; 3, Papillary Layer of Derm; 4, Angeiectases, occupied by Coagula and Blood-corpuscles; 5, Angeiectasis, with Dissepiment; 6, Oblique Section of Papilla.

subdivided by dissepiments, but it is probable that these partitions are really formed by the close juxtaposition of the walls of two or more dilated vessels. In a few places the angeiectases have been partially or completely obliterated by organisation of the contained blood-clot. (Fig. 3.)

It was noted that the spots became less distinct after the patient had been at rest for a few days in bed. No treatment for the condition was advised, and the man left the hospital after the healing of the surgical wound.